The Relationship Between Retinal Haemorrhages and Status Epilepticus and a Literature Review of Retinal Haemorrhages in Young Children

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Abstract

Objective: To examine the relationship between retinal haemorrhages and status epilepticus in children under two years of age. Methods: This study involved a retrospective chart review of patients up to 24 months with status epilepticus admitted to a regional hospital in Hong Kong from 1994 to 2002. Occurrence of possible preceding events that were associated with retinal haemorrhages was recorded. These included cardiopulmonary resuscitation, head trauma, eye infection, vomiting and severe coughing bouts, evidence of raised intracranial pressure, leukaemia, arteriovenous malformation and bleeding tendencies. Retinal findings, underlying cause of status epilepticus and radiological features were also reviewed. Results: During the study period, there were 23 children aged two years or below (mean 11.7 months old; range, 30 days to 24 months) admitted with status epilepticus. They all had a complete neurological and fundoscopic examination. Twenty-two (96%) children did not have retinal haemorrhage. The only child (4%) with retinal haemorrhage had evidence of non-accidental injury. Conclusion: This study suggests that status epilepticus per se is unlikely to be associated with retinal haemorrhage in young children. If retinal haemorrhage is present, one should be alerted to the possibility of non-accidental injury.

Key words

Retinal haemorrhages; Status epilepticus

Objective

To determine if retinal haemorrhages occur in children with status epilepticus in the local setting among the Chinese population.

Introduction

Retinal haemorrhage is frequently found in shaken baby syndrome. This is also the most common and familiar ocular sign of child abuse. Because this clinical sign is also associated with accidental injuries and a number of systemic illnesses, medical history is an important part of the clinical workup. The often unyielding history in cases of non-accidental injury necessitates a good understanding of the pathophysiology of retinal haemorrhage. The consequences of not diagnosing non-accidental injury and attributing retinal haemorrhages to other causes can be devastating for the child. With the increasing medical awareness and more understanding on the nature of retinal haemorrhage, early detection and appropriate intervention can be instituted so as to reduce further harm. We are unaware of any published study of fundoscopic examination.
findings of children who have status epilepticus. As patients with cerebral trauma may present with convulsion or status epilepticus, this study examines whether prolonged convulsion per se, as in status epilepticus, has any causal relationship with retinal haemorrhage.

Methods

This was a retrospective medical chart review of hospital admissions of all children up to two years of age diagnosed with status epilepticus whose clinical details could be retrieved from the hospital Medical Records Abstracting System since its inception in 1994 till December 2002. They all had a complete neurological and fundoscopic examination.

Status epilepticus was defined as generalised convulsion lasting 30 minutes or longer.

Information obtained included the demographic data, nature of seizure, any preceding events such as cardiopulmonary resuscitation, head trauma, infection involving the posterior segment of the eye, and bouts of vomiting and severe coughing. Underlying causes of status epilepticus, findings on ophthalmologic examination and radiological features were recorded.

Neonates with status epilepticus and non-convulsive cases were excluded from the study.

Results

Twenty-three children entered the analysis. There were a total of 16 males and 7 females. The age ranged from 30 days to 2 years with a mean age of 11.7 months.

Eight children (35%) were previously diagnosed with epilepsy, one child (4%) with Haemophilus influenzae meningitis, one child (4%) with pneumonia, seven children (30%) with atypical febrile convulsion, one child (4%) with inflicted head injury, one child (4%) with apparent life threatening event with hypoxic brain damage, one child (4%) with hypopituitarism and absent pituitary gland, one child (4%) with congenital adrenal hypoplasia and two children (9%) with no definite diagnoses.

Fundoscopy

Fundoscopic examination with direct ophthalmoscopy was performed on the 23 children by paediatrician. Twenty-two of them were normal and the one with abnormal findings was crossed checked by an ophthalmologist. Nineteen children were examined within 10 hours of the convulsion and the remaining four within 48 hours. It was not a routine practice to perform fundoscopy with mydriatics in all cases of status epilepticus if the fundal view was satisfactory.

Case of Non-accidental Injury

One child (4%) was found to have bilateral retinal haemorrhages. The retinal findings included bilateral dots and blots, intra-retinal, preretinal and vitreous haemorrhages. This child was a previously healthy 13-month-old boy who was admitted with status epilepticus. He was well when his mother left the house and entrusted him to the care of his father. His father admitted that he tossed him up and down to stop him crying. The father had one hand supporting the child’s neck and another on the back of trunk, with the child just leaving the father’s hands. It was also reported that he had a minor fall onto the floor while walking two days previously. There was no history of shaking of the child. The child developed status epilepticus with a deterioration of the neurological state. The history was incompatible with the clinical features. Together with the computed tomography finding of acute left subdural haemorrhage, this raised the concern for possible child abuse. A multidisciplinary team investigation concluded that he suffered from inflicted head injury. His fundoscopic examination was first performed by a paediatrician with direct ophthalmoscopy within one hour after admission. Retinal haemorrhages were found and they were confirmed by an ophthalmologist who used an indirect ophthalmoscopy method. His clotting profile was normal, with a platelet count of 323,000/mm³, prothrombin time of 10.9 seconds, and partial thromboplastin time of 20.9 seconds. He was blind with delayed global development and epilepsy on subsequent follow up.

Radiological Findings

Radiological evaluations were performed as clinically indicated. Nineteen patients (83%) had computed tomography scans of the brain. Out of these 19 children, four (17%) also had magnetic resonance imaging of the brain. Four patients (10%) had neither computed tomography scan nor magnetic resonance imaging of the brain. Fourteen patients (61%) had normal computed tomography scans. Five (22%) had abnormalities including
one with hydrocephalus from previous meningitis, one with suspicious encephalomalacia, two had hypoxic changes and one had acute subdural haemorrhage secondary to the inflicted brain injury.

Three of the four patients with MRI brain examination had abnormalities including one with absent pituitary gland, one with bilateral small infarct over lentiform nucleus and one with parietal subdural haemorrhage.

The subdural haemorrhage found in the CT and MRI brain scans were from the same child with extensive retinal haemorrhages. His clinical course was described in the section "case of non-accidental injury". Apart from the subdural haemorrhage, there was no additional MRI evidence of non-accidental injury.

Other Clinical Features

One child (4%) required cardiopulmonary resuscitation. One child (4%) had head trauma while cruising on the floor. Severe vomiting occurred in two children (9%). Severe coughing was recorded in two children (9%). None of the above patients had evidence of raised intracranial pressure, such as blurred disc, bradycardia, systemic hypertension, and they did not have any evidence of retinal haemorrhage. None of them had eye infection. The only patient with retinal haemorrhages was diagnosed with non-accidental injury. This child had head trauma while cruising on the floor. He had neither cardiopulmonary resuscitation, eye infection, severe vomiting nor coughing bouts.

Discussion

Retinal haemorrhages were reported in 5% to 23% of physically abused children. They may be unilateral or asymmetrical between the two eyes. In shaken baby syndrome, the incidence of retinal haemorrhages was reported to range from 53% to 80%. The incidence of bilateral haemorrhages is around 60%. The retina is a well vascularised multi-layered structure. Retinal haemorrhages are classified into pre-retinal (on the retinal surface), subretinal (underneath the retina) and intraretinal (within the retinal tissues proper) haemorrhages. Superficial intraretinal haemorrhage are known as flame shaped haemorrhages, where these are found within the nerve fibres. Deeper intraretinal haemorrhages are described as dot or blot haemorrhages when the haemorrhages are smaller or larger respectively. Haemorrhages involved most frequently the posterior pole in child abuse.

Apart from retinal haemorrhages, vitreous haemorrhages and traumatic retinoschisis can also occur. The appearance of vitreous haemorrhages can be delayed for two days to weeks after the shaking episode. Traumatic retinoschisis is the internal splitting of the retina.

To appreciate the association of retinal haemorrhages and shaken baby syndrome, the postulated mechanisms and other possible associations of shaken baby syndrome need to be discussed. The pathophysiology of retinal haemorrhages in shaken baby syndrome may be explained by several possible mechanisms. Firstly, the increased pressure transmitted to the central retinal vein from increased intrathoracic pressure, occurs as the perpetrator grasps the infant's chest. Secondly, direct trauma to the retina and retinal blood vessels results from repeated violent shaking, causing the vitreous to oscillate within the eyeball. Lastly, there is the implication of increased intracranial pressure impeding venous outflow from the eye, as when subarachnoid or subdural haemorrhage is followed by intraocular haemorrhages.

Other possible causes of retinal haemorrhages in infants include vaginal delivery, cardiopulmonary resuscitation (CPR), head injury from falling from height or motor vehicle accidents, haematological disorders, vascular malformations, meningitis, sepsis, hypertension, vasculitis and endocarditis.

The most common cause of retinal haemorrhages among infants is the process of delivery. They occurred in 19% to 23% of babies examined in the first 24 hours. The incidence is less in Caesarean section. They generally resolve by six weeks after birth. If they last beyond that period, one should consider the possibility of child abuse.

In a study on the occurrence of retinal haemorrhages in post cardiopulmonary resuscitation, Goetting found two out of 20 children after CPR had retinal haemorrhages. Kanter reported none of the candidates who underwent CPR had retinal haemorrhages when those who had co-existing severe systemic hypertension and head injury were excluded.

Falls are common in children. There have been a number of studies on retinal findings after head trauma. Johnson and Buys both described retinal haemorrhages occurring rarely in accidental head injury and the findings of such would suggest a non-accidental cause.

Amongst the few haematological disorders, childhood leukaemia is the most common cause of retinal
haemorrhages.\textsuperscript{19}

The presentation of cerebral vascular malformations, such as aneurysms when ruptured, closely resembled shaken baby syndrome.\textsuperscript{20}

It is most likely that the paediatrician is the first to examine the child. Paediatricians by training are more comfortable to work with an uncooperative child. Therefore a paediatrician experienced in managing child abuse can assess the victims, and consult an ophthalmologist if abnormalities are present during the ocular examination. Prompt recognition is needed since early detection may prevent further trauma from recurrent abuse.

In a review of 20 cases of shaken baby syndrome,\textsuperscript{7} Ludwig showed that shaken baby syndrome typically occurred in young children. Therefore all patients aged 24 months or younger were recruited in the current study. Two previous studies by Sandramouli et al\textsuperscript{23} and Tyagi et al\textsuperscript{24} showed none of their 64 children to have retinal haemorrhages following convulsions. Sandramouli et al concluded that convulsions rarely cause retinal haemorrhages and the finding of such warrants a detailed assessment to exclude non-accidental injury.\textsuperscript{23} Tyagi et al had a similar finding and they also recommended that the finding of retinal haemorrhages in a child with a history of convulsion should prompt further investigation to rule out other causes of these haemorrhages, particularly non-accidental injury.\textsuperscript{24} Mei-Zahav et al found retinal haemorrhages in less than 1% of patients who presented with convulsions, concluding that although theoretically convulsions can cause retinal haemorrhages in children, the prevalence is extremely low.\textsuperscript{25}

Theoretically, convulsions might potentially cause retinal haemorrhages in children as a result of a sudden rise in retinal venous pressure, secondary to an increase in intrathoracic pressure.\textsuperscript{3} Whether or not retinal haemorrhages are associated with status epilepticus may have medical implications.

In our study, we specifically looked at children with status epilepticus since previous studies only focused on convulsions in general, and the mean seizure duration was 10.5 minutes in one study.\textsuperscript{23-25}

In our study, 22 of the 23 children were found to have no retinal haemorrhages. The only one child with retinal haemorrhages seen within one hour was diagnosed to have abusive head injury after detailed social and medical evaluation and multidisciplinary case conference. In this child, the history was incompatible with the clinical features and the computed tomography finding of acute left subdural haemorrhage. The retinal haemorrhages were also very extensive. This raised the concern for possible child abuse. Despite the small number of patients in this study, the results of our current study are comparable to previous studies on convulsions.

The size of the study group is small as we only included those up to two years of age. The vascular system of this age group is more vulnerable to sudden changes in central venous pressure,\textsuperscript{1} rendering retinal haemorrhage more likely as in physical child abuse. Ideally, all patients should be seen by an ophthalmologist with mydriatics given as the false negative results are high in non dilated pupils examined with direct opthalmoscopy. In this retrospective study, all the patients except one were examined by a senior paediatrician with specialist qualification. A future prospective study involving the ophthalmologist with indirect ophthalmoscopy and the usage of mydriatics might be helpful in revealing more about the association of retinal haemorrhages and convulsion. Our review serves as a pilot study looking into this area.

**Conclusion**

We conclude that status epilepticus per se is unlikely to be associated with retinal haemorrhages in children under 2 years of age. Whereas it is difficult to establish the causal relationship of retinal haemorrhages and status epilepticus, our study revealed significant association between retinal haemorrhages and abusive head injury in the group of children presented with status epilepticus. Retinal haemorrhages, especially when extensive should alert all involved parties to look hard for any evidence of non-accidental injuries.

**References**

## Appendix

### Results of computed tomography and magnetic resonance imaging

<table>
<thead>
<tr>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Patient 3</th>
<th>Patient 4</th>
<th>Patient 5</th>
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<tbody>
<tr>
<td>Results of computed tomography</td>
<td>hydrocephalus from post meningitis</td>
<td>hypodense area adjacent to the temporal horn of the lateral ventricle in the temporal lobe. An oval shape CSF density lesion present medial to the stated hypodensity, suspicious of encephalomalacia</td>
<td>indistinct grey white differentiation, suspect early hypoxic changes</td>
<td>left parietal subdural haemorrhage</td>
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<tr>
<td>Results of magnetic resonance imaging</td>
<td></td>
<td></td>
<td>absent pituitary gland</td>
<td>left parietal subdural haemorrhage</td>
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CSF: Cerebrospinal fluid